

## Meniere's Disease

### **Overview**

Meniere's disease, or endolymphatic hydrops, is named after Prosper Meniere who was the first to describe the symptoms of fluctuating hearing loss, episodic vertigo, ear fullness, and ringing in the ear. The exact cause of Meniere's disease is not known, but we do know there is good evidence of increased fluid pressure of the endolymph portion of the inner ear. The episodic symptoms are most likely due to small breaks in membranes that allow the endolymph to mix with the perilymph. This causes fluctuating hearing loss and vertigo. The vertigo can last for minutes to hours until the membranes repair themselves and the fluids return to normal. The membranes are felt to repair themselves quickly and this explains why patients with Meniere's disease do not have continuous symptoms for more than hours at a time. With time the hearing loss may be progressive and can result in severe hearing loss with an inability to use hearing aids.

### **Diagnosis**

The diagnosis of Meniere's disease can be straightforward or rather difficult. "Definite" Meniere's disease is defined by the presence of documented fluctuating hearing loss (usually low-frequency), two or more episodes of vertigo lasting greater than 20 minutes, and ear fullness and/or ringing in the affected ear. Sometimes patients will have "probably" or "possible" Meniere's disease and will either not have the hearing loss or typical vertigo symptoms. In patients with only fluctuating hearing loss and no vertigo, a diagnosis of "endolymphatic hydrops" is made. Other conditions that may mimic Meniere's disease include migraine associated dizziness, vestibular neuritis, viral labyrinthitis, chronic subjective dizziness, or anxiety disorders.

### **Exam findings**

Exam findings are typically normal since the problem is in the inner ear. The bedside tuning fork exam may be abnormal. Some balance tests your doctor will perform may suggest balance weakness on the affected side.

### **Testing**

#### Audiogram

Multiple audiograms are essential for the diagnosis and to measure treatment response in patients with Meniere's disease. Usually the low-frequency hearing is abnormal, although the hearing loss can be seen as a flat or high-frequency pattern. Occasionally the hearing will return to normal when the disease is inactive.

#### Electrocochleography (ECoG)

ECoG is an office-based test where loud clicks are placed in the ear and a SP/AP ratio is calculated. If the ratio is greater than 0.37, increased inner ear fluid pressure is suggested. The ECoG test is not perfect and may be normal in patients with Meniere's disease and abnormal in patients with conditions that are not Meniere's disease. Because of these limitations, your doctor may or may not order this test.

#### Electronystagmography (ENG)

This is a comprehensive office-based balance test that looks at central balance function, positional balance function, and the inner ear reaction to warm and cold water. The findings are variable for patients with Meniere's disease. ENG is commonly obtained as a baseline before treatment and is used to measure residual inner ear function after treatment with gentamicin injections, labyrinthectomy, and vestibular nerve section.

### Vestibular Evoked Myogenic Potential (VEMP)

VEMP testing is a newer office-based test where clicks are placed in your ear and measurements are made from the large neck muscle called the sternocleidomastoid muscle. While VEMP testing is very useful in other conditions, it is limited in diagnosing Meniere's disease but may rule out other conditions.

Testing may be completely normal in patients with Meniere's disease, especially if the testing is performed between episodes of vertigo.

### **Treatment**

The first line of treatment is dietary management. A low salt diet is very important and will result in alleviation of the vertigo episodes in about 70% of patients. We recommend keeping the daily sodium intake to less than 1500 milligrams a day. It is important to not only avoid adding salt to your food but also to carefully look at nutrition labels, especially on soups, salad dressings, and sauces. Other items to avoid are chocolate and alcohol.

Patients that fail dietary management may start medication. The most common medication used in the United States is a diuretic. The diuretic decreases the pressure of the inner ear fluids. It is taken once daily and has few side effects. Another medication that is commonly used in Europe is betahistine. Betahistine is taken 2-3 times per day. It is only available through a compounding pharmacy. Sometimes other diuretics are used such as acetazolamide, but this commonly causes numbness around the lips and fingertips. Also, patients should avoid carbonated beverages while on acetazolamide.

In difficult cases, other medications may be prescribed. These are most commonly a benzodiazepine such as Klonopin or Valium. Other medications that may be helpful include Neurontin or amitriptyline.

If medical therapy fails, there are surgical treatment options available. These options depend on an individual's preference and depend on hearing level, severity of symptoms, and ability to live a normal life.

Treatments can be divided into ablative versus nonablative procedures. Ablative procedures include gentamicin intratympanic injections, labyrinthectomy, and vestibular nerve section. The results of these interventions are irreversible and lead to permanent destruction of the inner ear balance system. This could be important if you develop Meniere's disease in the other ear, which can occur up to 10% of the time. Nonablative procedures include intratympanic steroid injection, Meniett device, or endolymphatic shunt surgery. These interventions usually preserve inner ear balance function and rarely cause more hearing loss.

### Intratympanic steroid injection (IT steroids)

IT steroids are a safe and effective treatment for patients with Meniere's disease. A study from Johns Hopkins found 91% of patients who underwent IT steroid injections after failing medical management were vertigo free with a two year follow up. Many of the patients required multiple injections, but the procedure is well tolerated and is done on the clinic.

The procedure is performed in the office and is well tolerated. The ear drum is numbed using a variety of methods. A small amount of solution with steroids is injected through the ear drum into the middle ear space. The steroids go through the round window of the inner ear and into the cochlea. You then lay on your back for 20 minutes. There is a 3 in a 1000 chance you could develop a hole at the injection site. Otherwise the risks are low, and you can safely have multiple injections.

#### Intratympanic gentamicin injection (IT gentamicin)

IT gentamicin is given the same way as the IT steroids; however, gentamicin destroys the balance cells. This makes this procedure an ablative procedure and once the balance cells are gone, there is nothing that can be done to restore them. There is also a chance of further hearing loss with gentamicin since the gentamicin may also damage the hearing cells. Patients often complain of constant imbalance for 1-4 weeks after an injection. Risks of the procedure are a hole in the ear drum that may need repaired, hearing loss that could result in deafness in the treated ear, and need for multiple injections that sometimes do not work.

#### Meniett device

The Meniett device is a low-pressure pulse generator you wear for about 5 minutes at a time 3 times a day. The exact way it works is unknown, but it is felt it may help push the inner ear fluids and increase absorption of the fluid resulting in less fluid pressure. An ear tube (like children get when they have ear infections) has to be placed and has the risk of hearing loss, hole in the ear drum that may need repaired, infections and drainage, or the need for removal of the tube that is in for too long. The device can be difficult to obtain through some insurance companies, so it takes time for insurance approval.

#### Endolymphatic shunt/decompression surgery (ELS)

ELS is a nonablative surgical option. Under general anesthesia, a small incision is made behind the ear. The mastoid bone (bone under the ear) is removed and the endolymphatic sac is visualized. Your surgeon may either decide to just remove the bone around the sac or to place a small piece of plastic (shunt) in the sac. About 70% of patients that fail medical management report decrease in vertigo episodes after ELS. The surgery takes 1-2 hours and is day surgery. The risks include hearing loss including total hearing loss, facial paralysis, chronic dizziness, change in taste, spinal fluid leak, meningitis, failure to relieve symptoms and need for further procedures.

#### Vestibular nerve section (VNS)

The VNS is successful at relieving vertigo in about 95% of patients. The major drawback is you have to undergo a craniotomy. The procedure lasts 2-3 hours, but requires an overnight stay in the ICU and 2-3 more days in the hospital. Patients usually have significant dizziness that may require outpatient balance therapy. Hearing loss is not common but is more common than with IT steroids or the ELS surgery. Risks include bleeding that could rarely be life-threatening, meningitis, spinal fluid leak, chronic headaches, chronic dizziness, facial paralysis, and need for further procedures.

#### Labyrinthectomy

A labyrinthectomy is also 95% successful in alleviating vertigo. Patients who undergo a labyrinthectomy lose all residual hearing, so this procedure is performed only for patients with poor hearing. The procedure is through the same incision as the ELS surgery and a mastoidectomy is performed. It is common for patients to have significant dizziness after surgery and some patients will require outpatient

balance therapy. Risks of surgery include facial paralysis, spinal fluid leak, meningitis, and failure to control vertigo.

### **Hearing loss and ringing in the ear (tinnitus)**

You may have noticed the treatment options were most successful at controlling the vertigo symptoms. The hearing loss can be progressive despite treatment. Sometimes hearing aids are the best option; however, the hearing loss can be severe enough hearing aids are not useful. If the hearing loss is this severe, there are options you can discuss with your doctor or audiologist.

Tinnitus can be very bothersome. The best treatment is avoidance of quiet situations including using something to generate a background noise while sleeping. If indicated, hearing aids can decrease the severity of the tinnitus. Occasionally the tinnitus is so severe that medications or other therapy may be required.

### **Conclusions**

Meniere's disease can be disabling and make leading a normal life difficult. We rarely have a patient we can't make significantly better or symptom free by using the treatments outlined here.